Critical Digest.

THE PATHOLOGY OF SYRINGOMYELIA.

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There is at present little discussion regarding the symptomatology or diagnosis of syringomyelia. The disease is readily recognized, and many cases have been recorded in which the diagnosis has been confirmed by post mortem examination.[1] The extensive literature of the subject is to be found in Schlesinger's[7] and Dimitroff's[8] monographs, where more than five hundred references to published articles are given.

The chief interest in the disease now centres about its pathology, regarding which many diverse views prevail. This diversity of opinion is traced by Hoffmann[1] to the fact that the cases which have been examined have rarely if ever come to a fatal end in the early stages of the disease, and hence the origin of the process has to be deduced from the terminal appearances. These appearances are subject to individual interpretation, and each pathologist in turn has tried to bring all cases into conformity with his own observations upon his own particular cases. The various views which have been brought forward to explain the existence of cavities within the spinal cord are, in the order of their proposal, as follows:—

a. The view of Virchow, who ascribed all such cavities to a distension of the central canal. This view was modified by Leyden, who considered the condition a congenital hydromyelus. These views are now modified by their authors.

b. Simon first distinguished sharply between hydromyelus and syringomyelia, and pointed out the existence in the latter of gliomatous tissue and its destruction.

c. Schultze then urged the view that prior to the formation of any cavity there was always a central gliomatosis, a proliferation of glia cells, which, though usually beginning near the central canal, may begin anywhere in the cord.

d. Hallopeau later proposed the theory that the condition was really a chronic inflammation beginning in the epithelium of the central canal or in the periependimal cells, and considered the process of the nature of sclerosis.

e. Joffroy, like Schultze, denied any necessary relation between the pathological process and the epithelium of the central canal, but considered that an inflammatory change might begin in any part, leading finally to disintegration, a true "myelite cavitaire."

f. Langhaus then brought forward an hypothesis, which Kronthal subsequently supported and which Müller and Medin[6] have recently urged, that an obstruction to the venous or lymphatic return flow from the cord may lead to accumulation of fluid in the central canal, distension of the cavity of the canal, pressure on, and thus disintegration of adjacent gray matter, with glia formation.

g. Lastly, Van Gieson, from a study of a number of cases, has shown that a long cavity may be left after the absorption of the clot formed by a perforating hemorrhage in the cord; that such a cavity is surrounded by a zone of neuroglia, thus simulating true syringomyelia. Dana, while admitting the possibility of perforating hemorrhage confined to the gray matter of the cord, considers that a true necrotic process is in some cases the primary cause of such cavities as Van Gieson describes.

When the various views regarding the exact origin of cavities in the cord are tabulated, it appears that the following views have a well-established basis in pathological observation :—

Cavities may be produced by

I. A dilation of the central canal.
  a. Congenital from maldevelopment.
  b. Acquired as hydrocephalus.
II. A maldevelopment of the central canal.
  a. With supernumerary canal or double canal.
  b. With imperfect closure of the posterior septum.

These conditions (one or more) are thought by many authors to be present as an original predisposing cause in every case of syringomyelia. It is to be remembered that the central canal is surrounded by a large number of small epiblastic cells, whence the neuroglia developes in a normal cord, and that the formation of new neuroglia tissue is a part of the lesion in syringomyelia, and that the situation of the cavity or of the neuroglia proliferation. points to some necessary relation between these epiblastic elements and the new tissue.

III. A primary proliferation of the neuroglia tissue of the cord, without any maldevelopment, and a gradually advancing disintegration of the new tissue at its centre, may lead to the formation of a cavity with walls of thick neuroglia. This is termed gliomatosis.

a. This may be a single hyperplasia from unknown causes.

b. It may be inflammatory, produced by trauma, by irritation of poisons, or by infection.

c. It may be a true tumor formation.

There may be a relation between the existence of numerous embryonal cells, capable of development into neuroglia, near the central canal, and the process of gliomatosis, as it seems reasonable that such a process should begin in these cells when from any cause it is set up. But authors dispute this relation, and some hold that the proliferation may begin anywhere in the cord, in normal neuroglia tissue. In this connection the views of Weigert regarding neuroglia may be cited, and his opinion concerning the lesion in syringomyelia.

Weigert (Die Neuroglia, 1896) shows that new formations of neuroglia may be either cellular in structure or fibrous in structure. If cellular, the new formation is a true glioma, such as is found in tumors of the brain or spinal cord, and in such a glioma fibres are few. Weigert calls attention to the fact that this is not the structure of the glia tissue about the cavity of syringomyelia, but that the neuroglia found around this cavity consists almost exclusively of glia fibres with few cells, and that these fibres, though extending in all directions, are chiefly vertical in their course. Miura[4] also has shown the sharp contrast between ordinary glioma, even glioma containing a cavity, and the gliomatous condition of the cord in syringomyelia. Weigert holds that the neuroglia is merely a substance produced by nature to take the place of nerve tissue which has been destroyed, and that its proliferation is always an indication that the nerve tissue has primarily disintegrated. Some destruction of nerve tissue would therefore, according to his idea, precede the formation of gliomatous tissue, and hence he discards wholly the theory of syringomyelia to which the term spinal gliosis has been applied. Weigert says "many authors believe that the essential lesion in syringomyelia is the formation of a tumor followed by softening and the formation of a cavity. It is admitted that there is a growth of neuroglia of the typical fibre-type about the cavity. But this fibre mass, devoid of cells, does not resemble a glioma, and there is no reason to believe from the mere presence of neuroglia that the cavity is not a congenital or acquired abnormality of the central canal. There is a thick cluster of neuroglia fibres normally about the canal. By the pressure in this enlarged canal the nervous tissue may be destroyed and hence a growth of neuroglia fostered. If the pressure increases the neuroglia may also be destroyed, and in its place about the cavity a hyaline formless mass may remain. The gliosis is not the essential feature; it is only a secondary result." (Centralbl. f. Path. u. Path. Anat., Vol. I., p. 736. and Neuroglia, p. 101.)

On the other hand, many authors have noticed the great proliferation of neuroglia cells in the segments of the spinal cord above and below the limits of the cavity and have considered this a proof that the proliferation of these cells was a necessary precursor to the formation of the cavity, a view exactly opposed to that of Weigert.

Some authors (Miura) have sought to contrast the true glioma sometimes found in the cord with the gliomatosis of syringomyelia, but Turner and Mackintosh[9] have published cases in which every possible transition between these conditions is evident. It is true that sometimes a glioma, or a sarcoma, or a gliosarcoma has been found within the cavity (Van Gieson) in cases of syringomyelia, and Hoffmann[1] describes these cases as a special class. But it is not always possible to determine whether the cavity or the tumor is the primary condition or whether both were varieties of the same pathological process going on in different ways at different levels of the cord. Miura has described a case in which there was a diffuse glia proliferation in the gray and white matter from the mid-cervical to the upper lumbar region without any cavity formation whatever, but with here and there pale foci which might be considered areas of beginning degeneration. This he considers a true tumor formation, not a chronic inflammatory neuroglia proliferation and without any relation to the central canal. In Turner's cases the tumor and the canal were not at the same level, but ran into one another, and here, also, there was no evident relation between the canal and the pathological processes.

IV. A softening and disintegration of cord tissue may be followed by the formation of a cavity with a wall of new neuroglia or without any wall. Such cavities are thought to be due to disease in the walls of the spinal arteries. Müller and Medin[6] have seen a cavity in the gray matter of the cord with walls of normal nerve tissue with no signs of inflammatory disease or of proliferation in the glia, the size of the cavity corresponding in situation to the degree of endarteritis in the spinal vessels, and having no definite relation to the central canal. Wieting[7] has described a cord containing numerous cavities, due entirely to the low nutrition of the nerve tissues and consequent necrosis from disease of the spinal arteries in connection with meningomyelitis. The cavities were of various shapes, and their walls contained disintegrated neuroglia cells and many blood vessels. Here and there the wall of the cavity was made of thick neuroglia with many fibres and numerous cells. At other places a hyaline infiltration was seen in the wall of the cavity and in the surrounding tissue, which was considered a terminal phase of glia disintegration. In this cord many collections of neuroglia cells in foci and stripes along the blood vessels were seen. There was no relation between the situation of these cavities and the central canal. The capillary vessels were increased in the foci of neuroglia and in the walls of the cavities forming a tangled mass with thick sclerotic walls and sometimes with obliterated lumen. These appearances do not, however, resemble at all closely the appearances characteristic of syringomyelia, and Hoffmann rightly discards such cases of necrotic spinal cavities from the classes of pathological varieties of syringomyelia. It has been thought by Kronthal that lymphstasis within the cord and consequent necrosis produced by transverse compression of the cord may lead to the formation of cavities which may or may not communicate with the central canal, but this theory is doubted since compression of the cord by tumors or after Pott's disease is not found to cause cavities, and such cavities as he describes certainly differ from those found in syringomyelia in the absence of a wall.

V. Lastly, there is not wanting a theory of the disease which would trace it to an infection of the cord by a bacillus. The resemblance between Morvan's disease and syringomyelia has been noticed by all writers, and their identity has at last been established by the fact that in more than a dozen cases of Morvan's disease the lesion of syringomelia has been found post mortem in the spinal cord.[1] It is true that in Morvan's disease an infection of the peripheral nerves has also been found to exist in some cases, but it remains an open question whether this peripheral neuritis is not an expression of a disturbance of trophic action due to the lesion in the spinal cord. Hoffman considers that the cases of Morvan's disease are merely a variety of syringomyelia and maintains that there are no clinical differences between the affections. But the similarity between Morvan's disease and lepra anæsthetica has been noticed by more than one author, Zambaco having especially called attention to the resemblance. It is known that lepra anæsthetica is produced by a bacillus, and many writers upon Morvan's disease have maintained that the fact that this disease develops, particularly along the coast of Brittany, among fishermen, and in certain localities rather than in others, offers a positive proof that it has some extraneous origin, probably of the nature of an infection. In a very exhaustive article on the relation between syringomelia, lepra and Morvan's disease, Prus[11] reaches the conclusion that these three diseases may be varieties of the same general condition, Morvan's disease being the least intense, lepra anæsthetica the most severe. Lepra depends upon the existence of a bacillus which is now well recognized. Strauss has found this bacillus under the skin in cases of Morvan's disease which he examined in Brittany, and this has been confirmed by Moniez and Roux. Prus, therefore, has proposed the hypothesis that the lepra bacilli, after penetrating the skin, may set up a degeneration of the nerve fibres, causing the well-known symptoms of Morvan's disease; that then they may migrate along the course of the nerves, leading to changes in the nerve trunks and thus ascending the nerves to the spinal cord, where they may produce gliomatosis, with its subsequent destruction, and hence the appearance of syringomyelia. Prus does not affirm that all cases of syringomyelia originate in this manner, but believes that a certain number may be thus explained. On the other hand, it is to be remembered that Schultze draws a sharp line between lepra and syringomelia and denies the existence of any spinal lesions in lepra. And other authors who have examined the spinal cord in lepra have found the bacilli of the disease in the cord and in the cerebro spinal fluid without finding any condition of gliosis or any degeneration of the nerve tissues. The identity of lepra and syringomelia is by no means established, and therefore this theory of an infective cause for syringomyelia cannot be as yet accepted.

The conclusion to be drawn from this review of various theories is that cavities develop from many causes, and that the lesion of syringomyelia is not of uniform causation.

[1] Prof. J. Hoffmann. Zur Lehre von der Syringomyelie. Deut. Zeitschr. f. Nervenheilkunde, III., 1893.

[2] Prof. A. Chiari. Ueber die Pathogenese der sogennanten Syringomyelie. Zeit. f. Heilkunde, IX., 307.

[3] Joffroy et Achard. Syringomelie et Maladie de Morvan. Archiv de Med. Experimentale et d'Anatomie pathologique, III., annee 1.

[4] A Miura. Ueber Gliom des Rückenmarkes und Syringomyelie. Ziegler's Beiträge zur Path. Anat., XI , 91, 1892.

[5] F. Wieting Ueber einen Fall von Meningo-myelitis Chronica mit Syringomyelie. Ziegler's Beiträge zur Path. Anat., XIX., 215, 1896.

[3] Mueller und Mendin. Zur Kenntniss der Syringomyelie, Zeitschr. f. Klin. Med.. XXVIII., 128, 1895.

[7] H. Schlesinger. Die Syringomyelie. Leipzig, 1895.

[8] S. Dimitroff. Uber Syringomyelie. Archiv. für Psych., XVIII., 1896.

[9] Turner and Mackintosh. Three cases of new growth with cavity formation in the Spinal Cord. Brain, Pt. LXXV., 1896.

[10] Babes et Manicatide. Recherches sur la Syringomyelie. Archives dep Sci. Med. de Bucharest, May, 1896.

[11] J. Prus. Die Morvan'sche Krankheit, ihr Verhältniss zur Syringomyelie und Lepra. Archiv. für Psych., XXVI., 771, Nov., 1895.